Eosinophilic Granuloma of Bone: *
Report of Nine Cases

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THE TERM *eosinophilic granuloma* was first used in 1940, by Lichtenstein and Jaffee.[7] In the same journal but submitted about a week earlier, Otani and Ehrlich[10] published an article describing this lesion under the title "Solitary Granuloma of Bone." The lesion had been described earlier by Mignon[8] and Finzi[3] but it was believed to be due either to myeloma or osteomyelitis.

The first cases reported were examples of solitary bone lesions. In 1942, Green and Farber[4] reported ten cases of eosinophilic granuloma of bone some of which were multiple lesions. They pointed out that eosinophilic granuloma of bone, Hand-Schüller-Christian's disease, and Letterer-Siwe's disease are different manifestations of the same pathological process. More recently, Arnold,[1] and Weinstein *et al.*[13] have reported eosinophilic granuloma of bone in association with extra-osseous lesions, particularly in the lung parenchyma. According to O'Neill *et al.*,[9] 187 cases of eosinophilic granuloma of bone were recorded in the literature through 1954. Since that time many more cases have been reported.

In 1953, Lichtenstein proposed the term histiocytosis X to describe the three manifestations of reticulosis, for he believed the pathologic common denominator was a specific inflammatory histiocytosis. Eosinophilic granuloma is a localized form of histiocytosis X. Disseminated histiocytosis X is divided into an acute or subacute form (Letterer-Siwes's disease) and a chronic form (Hand-Schüller-Christian disease). Eosinophilic granuloma may have lung or other visceral involvement in addition to bone lesions, and indeed, primary pulmonary eosinophilic granuloma has been reported in the literature in several instances.[11] There is not a clear-cut separation of the three forms of this disease so that in many instances it may be difficult to know where to classify a given case. Many authors believe some cases may change from the localized to the disseminated form. It does seem justifiable on clinical grounds, however, to make the separation into three divisions; this presentation is concerned only with manifestations of eosinophilic granuloma of bone.

Eosinophilic granuloma of bone is a disease of children and young adults. According to Teplick and Broder[12] only 5.0 per cent occur in persons over 30 years of age and 75 per cent of the lesions are solitary. They also state that 59 per cent of the lesions involve the skull, ribs and femurs, although any bone may be involved. O'Neill *et al.*,[9] in their review of 189 patients found that 119 of the cases were males and 48 females. In 22 patients sex was not listed. Pulmonary involvement in the form of miliary infiltrations throughout both lungs may occur. There is usually a history of pain and swelling over bony lesions and often localized tenderness. There may be slight fever, leukocytosis with eosinophilia and increased sedimentation rate.

The roentgenologic appearance of the skull lesions is manifested by sharply demarcated areas of destruction of any size with irregularly scalloped borders. Usually both tables of the skull are involved and there is no periosteal reaction. Lesions in the pelvis, scapula, and sternum are similar

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TABLE 1. *Summary of Cases*

Patient	Sex Age	Duration of Symptoms	WBC	Sedi. Rate	Location	Treatment*
1. D. T.	M 14	13 months	9,100	17	Acetabular roof	Curettage
2. C. T.	F 13	5 weeks	9,600	32	Femoral shaft	Curettage
3. E. M.	F 3	3 weeks	14,200	53	Fifth lumbar body	Curettage
4. W. J.	M 9	Pathologic fracture	5,600	—	Humeral shaft	Bone chips and cast
5. J. P.	M 8	4 weeks	8,200	—	Tibia	Curettage
6. K. W.	M 12	2 weeks	5,050	17	Sternum	Curettage
7. S. S.	F 9	8 weeks	9,600	32	Femur	Curettage and roentgen therapy
8. S. G.	F 2	6 weeks	9,200	—	Skull (parietal)	Curettage
9. G. D.	M 8	4 weeks	9,900	—	Skull (parietal)	Curettage and roentgen therapy

* Lesion healed in all instances.

to those in the skull. In the long bones, there is usually expansion of the shaft sometimes with periosteal new bone formation and a central area of medullary bone destruction. Pathologic fractures may occur. Lesions in the long bones seldom occur below the elbows and knees and seldom involve the epiphyses. In the mandible there are areas of destruction around the teeth so that the teeth appear to be floating in the mandible. The differential diagnosis of the skull lesions includes myeloma, epidermoid cyst, metastasis and osteomyelitis. In the long bones the differential diagnosis includes Ewing's tumor, osteomyelitis, fibrous dysplasia, metastasis, myeloma and giant cell tumor.

Pathologically the lesions consist of fibrous stroma with histiocytes, giant cells and eosinophils and occasionally foam cells. Histiocytosis is an inflammatory proliferative process. The cause is unknown and cultures have been reported negative by most authors.

To establish an accurate diagnosis, it is believed that all of these lesions should be biopsied. At the time of biopsy the soft gray material which occupies the area of bone destruction may be curetted. This may be sufficient to cause regression of the lesion. If multiple lesions exist, it is necessary to biopsy only one of them and the remainder may be treated with roentgen therapy. The recommended dose of roentgen therapy is a total of 600 to 800 r to the lesion in two or three treatments over a period of two to four weeks. With curettage or roentgen therapy the lesions begin to regress in a few weeks and should be completely healed in a few months. Without therapy regression of the lesions is much slower. Eosinophilic granuloma of bone, however, is a benign lesion and the outlook for complete recovery is excellent. A few of the cases may progress into the more serious and chronic form of Hand-Schüller-Christian disease.

We are reporting nine cases of solitary eosinophilic granuloma of bone. All of the cases were proven by biopsy (Table 1).

Case Discussion

In Case 1 the lesion was in the acetabular roof and the symptoms had persisted for over a year before the diagnosis was made. An x-ray

of the pelvis when the symptoms first started was negative. Adequate exposure of this lesion was obtained by a posterior approach to the hip joint.

Case 2 showed an obvious lesion of the femoral shaft but the periosteal reaction suggested the onion skin appearance of Ewing's tumor which has frequently been mentioned in the literature.

Case 3 involved the body of the fifth lumbar vertebra. The symptoms were of short duration but the pain was so severe that the child refused to walk. The lesion was suspected on plain x-rays of the spine, but was clearly shown as a large area of destruction by body section roentgenography. An anterior approach to the vertebral body was necessary to curette the lesion. Although curettement was incomplete the relief of pain was noticed when the patient awoke from the anesthetic and x-ray examination six months later showed the lesion to be almost completely healed. Compere *et al.*[2] have shown that vertebra plana may result from eosinophilic granuloma involving a vertebral body.

Pathologic fractures through these lesions are uncommon. In 1949, Hill [5] reported four cases in a review of the literature. Case 4 (Fig. 1) had no symptoms or treatment before the fracture occurred. It healed without incident.

Case 5 (Fig. 2) was a lesion of the upper tibia. The skin over the lesion was red, warm and tender. This reaction subsided following curettement and wound healing.

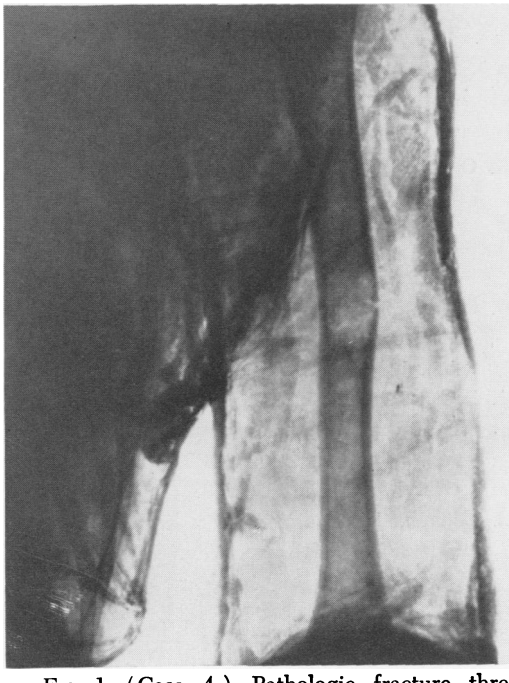

FIG. 1. (Case 4.) Pathologic fracture three weeks after curettage. The fracture line and lesion can be seen through the cast.

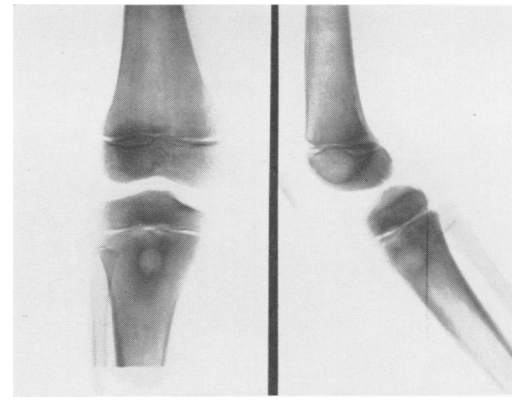

FIG. 2. (Case 5.) The lesion in the proximal portion of the tibia shows central destruction with surrounding bone reaction.

Case 6 was brought to attention following minor trauma to the sternum. The lesion had broken through the cortex of two segments of the gladiolus of the sternum. It healed following curettement.

Case 7 (Fig. 3) was another lesion of the femoral shaft which resembled Ewing's tumor. The patient walked with a limp and the thigh was swollen and tender. The lesion healed following curettement and roentgen therapy of 800 r to the lesion in six weeks.

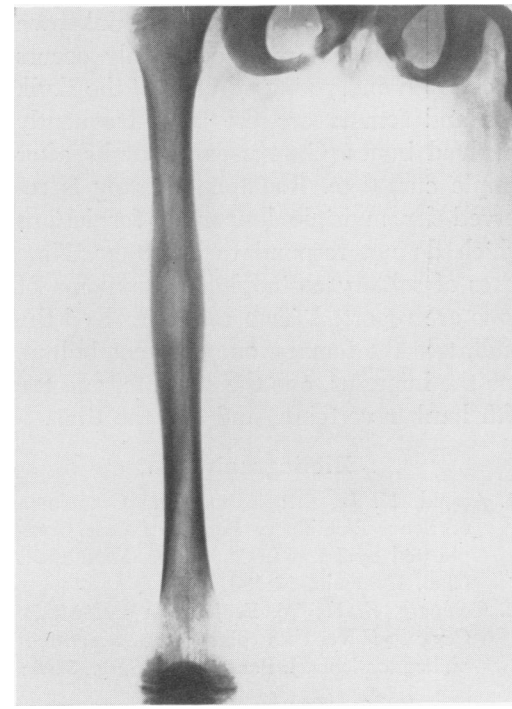

FIG. 3. (Case 7.) Cortical bone thickening and periosteal bone proliferation can be seen around the lesion in the femur.

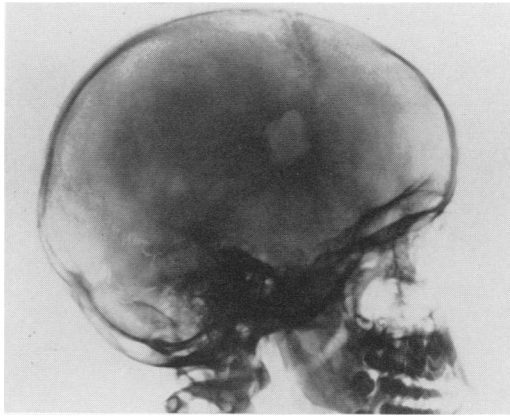

FIG. 4. (Case 9.) The typical skull lesion is seen in the parietal bone.

Case 8 was a typical lesion involving the parietal bone of the skull. It was treated by curettement alone and there was still evidence of its presence on a skull x-ray 18 months later. Five years later, however, the skull x-ray was negative.

Case 9 (Fig. 4) was also a typical skull lesion involving the parietal bone. It was treated by curettage and roentgen therapy and four months later (Fig. 5) the lesion was completely healed.

Summary

Eosinophilic granuloma of bone is a benign inflammatory reaction to an unknown etiologic agent. It most commonly occurs in children and young adults and the skull, ribs and femurs are the most frequently involved bones. Cure usually results after simple curettage. Roentgen therapy is reserved for multiple lesions or for lesions which do not respond to curettage. Nine cases of solitary eosinophilic granuloma of bone are reported. Two cases involved the skull, two the femurs, one the acetabulum, one the humerus, one the sternum, one the fifth lumbar vertebra, and one the tibia.

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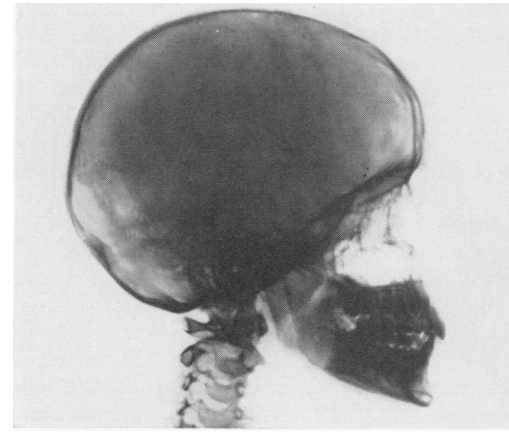

FIG. 5. (Case 9.) Four months after biopsy and roentgen therapy. The skull is normal.

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